HEREDITARY CEREBELLAR ATAXIA AND GENERAL PARESIS

A Supplementary Report and a Correction.

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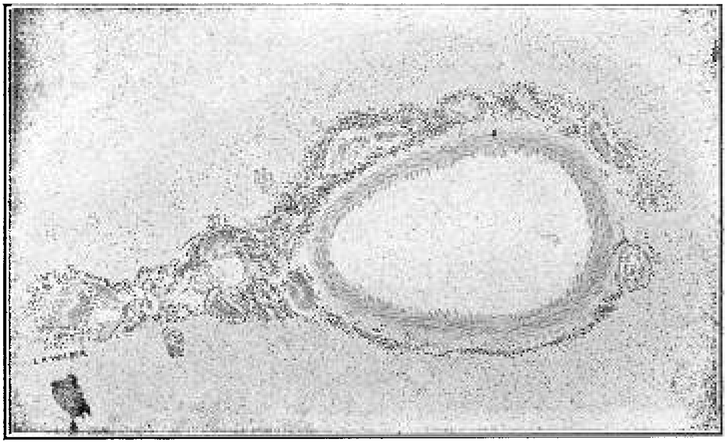
In the Journal of Nervous and Mental Disease, March 1902, I published a paper entitled Hereditary Cerebellar Ataxia with Report of a Case. The title was one of expediency. I quote the last paragraph preceding the report.

"Doubting the existence of a disease meriting the title hereditary cerebellar ataxia, I have still for convenience called my case by this name. Should there really be such a malady, I am naturally in doubt as to whether I am reporting an example of it."

My case was that of a young man eighteen years old when first seen, who presented many of the symptoms described by Marie as belonging to hereditary cerebellar ataxia, with very considerable mental failure added. I quote a paragraph following the report.

"In this case the progressive mental failure, rigid pupils, ataxic gait with exaggerated reflexes and indistinct speech, together with the history of miscarriage and death in infancy, at once suggest precocious general paresis due to inherited syphilis. On this supposition the elder brother may have had the same disease, or, bearing in mind the hemiplegia, *syphilis hereditaria tarda.* Remembering that precocious paresis is apt to be atypical, I have had this possibility constantly in mind. Even now I have no inclination to be dogmatic on the subject, but the case has never looked to me like general paresis, and I think that diagnosis could not be maintained. Adequate evidence of syphilis in the parents is wanting. What Freud noticed in the family of his diplegics and called *eine Neigung zur Leichtsterblichkeit* (vital vulnerability) in my opinion explains the early deaths and miscarriages in the family as well as does the theory of specific disease. Incoordination distinctly antedated mental deterioration, and has throughout dominated the clinical picture. No trace of a delusion has ever been detected; tremor of lips, tongue and hands has been consistently absent. In addition, absence of the Argyll-Robertson pupil, of analgesia of the legs, of good-natured self-satisfaction and mental depression alike, would tend to exclude paretic dementia. Extreme over-action of the facial muscles is no part of the symptomatology of this disease, and by this time there should be more motor feebleness than is the case."

The report carried the patient almost to the time of death. He became entirely helpless, quite demented, greatly emaciated and died of marasmus. The brain and spinal cord were given to Dr. Lewellys F. Barker, and his findings seem to show conclusively that I was entirely wrong and that the case was one of general paresis, probably due to inherited syphilis.



Showing Cellular Infiltration of the Pia and About a Blood Vessel.

The following is Doctor Barker's report, and I am also indebted to him for the excellent sketch herewith produced. In a note accompanying the report he says: "I suppose there might be a difference of view as to whether this is 'paretic' or simply 'syphilitic' but I regard it as paretic."

Report on the Pathological Anatomy of Dr. Patrick's case.

Pia Mater Cerebri. Macroscopically turbid and thickened. Microscopically everywhere infiltrated with lymphoid cells, especially around the blood vessels, but also diffusely among the connective tissue strands. The vessels are distended with blood, the whole pia is thickened not only on the surface of the gyri, but in the depths of the tissues. Wherever the pia extends the aggregations of lymphoid cells are met with. In the accompanying figure, the infiltration (especially perivascular) with small mononuclear cells in a fold of the pia extending deep into a sulcus is well shown.

Cortex cerebri. The capillaries and small blood vessels are distended with blood and are prominent in the microscopic picture. The walls of a great many vessels show aggregations of lymphoid cells in the perivascular spaces.

The nerve cells of the cortex are smaller than normal, many of them being distinctly shrunken. There is marked proliferation of the glia and in hematoxylin or separation glia nuclei stand out prominently. In Weigert preparations the fine medullated fibres of the cortex are decreased in number.

Spinal cord. Marchi and Weigert preparations reveal no degeneration in the white matter. The gray matter looks normal. The cord is large and well developed. The only abnormality met with is an occasional group of lymphoid cells in the pia, but the pia mater spinalis is nowhere invaded with these cells as the pia mater cerebri is. Sections were cut from every segment of the cord.

Cerebellum. Section were made through both hemispheres and through the vermis. The cerebellum is large and well formed. No sclerosis. No atrophy. The cerebellar nuclei show no alteration.

Pathological-Anatomical Diagnosis. Chronic meningo-encephalitis.

LEWELLYS F. BARKER

Papers published since mine have rendered it almost devoid of interest but this addendum and correction are due those who may have read it and to Dr. Barker who made such a painstaking examination of the material.